Pain and disabilities related to hip disorders in adults with severe cerebral palsy

Boldingh, E.J.K.

2013

document version
Publisher's PDF, also known as Version of record

Summary

Chapter 1 of this thesis presents an introduction and outline of the thesis, and explains the rationale for the research. While working as a physiatrist for children, I was confronted with the question: are patients with cerebral palsy (CP) who develop hip disorders bothered by this disorder, in their youth, in adulthood and / or later? And if so, how can we help these people by preventing or curing this problem?

The chapter reports about the development of the research design and a suitable instrument to measure pain in the patients of the study sample. After the study among 160 patients with CP level V (in the GMFCS classification) had been completed, a systematic review of the literature about preventive, corrective and palliative surgery was set up.

The results of the patient study and the systematic review enabled us to develop a decision tree for the treatment of hip disorders in spastic cerebral palsy. The decision tree was presented and discussed at the regular meetings of Dutch pediatric physiatrists, and is now considered to be the Dutch consensus.

Chapter 2 describes the development of a pain assessment instrument (PAICP) for patients with cerebral palsy, which was then tested in a cross-sectional validation study. Measurements were made at homes for the severely handicapped and in the home environment of the patients. The participants were 164 adult patients with severe CP, their caregivers and physiotherapists. The study was preceded by a pilot study with nine healthy children.

The PAICP instrument uses drawings of everyday situations. Patients score the pain they experience on a faces pain scale. Reproducibility and construct validity were assessed in a pilot study with CP patients and healthy children. The construct validity and agreement between the pain scores of the patients and proxies was assessed in 160 patients with severe CP. The main outcome measure was the pain score on PAICP.

The results showed that the instrument had good construct validity and sufficient test-retest reproducibility. A significant difference was found between the mean scores for 'painful' and 'non-painful' situations. We found moderate agreement between the scores of the patients and the proxies regarding daily activities, and only for those activities in which the proxies were personally involved. There was significant agreement in the scores of the patients and their proxies in hip straining situations.

We concluded that the PAICP has satisfactory test-retest reproducibility and construct validity. It provides an indication of the pain experienced by patients in situations in

which proxies are not personally involved and may also be more valid than proxy measures for other situations.

A striking finding was that stretching of the legs by physiotherapists, a common procedure in this group of patients, is actually a painful experience for them. Further research is needed to evaluate the benefit of the physiotherapy these patients receive to prevent or treat the joint contractures they all develop.

In **chapter 3** we report on a study of the relationship between migration and deformity of the femoral head and osteoarthritis on the one hand, and pain on the other hand, in people with severe CP. Measurement instruments that enable the patients themselves to indicate the severity of their pain had not previously been used to study this relationship.

A cross-sectional study was carried out to investigate the relationship between hip X-ray findings (migration, deformity and osteoarthritis) and pain in 160 adults with severe CP. The patients were unable to walk independently (GMFCS level V), and had a minimum mental age of four years. They scored their pain on the PAICP instrument. Their scores ranged from 1–7. Pain was dichotomized, and logistic regression analysis was performed to evaluate the associations between X-ray findings and hip pain.

Thirty patients out of a total of 160 (19%) had hip pain in hip-straining situations. If an adult with GMFCS level V has a hip disorder, this increases the risk of pain from a basic level of about 10% – not related to hip disorder – to a risk associated with hip disorder of about 30%. Severe deformity of a femoral head was present in 41 (30%) patients. Osteoarthritis was found in 23 (16%) patients. Twenty patients who had an abnormal configuration of the femur and pelvis, mainly after (unsuccessful) bone surgery (n=17, 85%), were analyzed as a separate group. In this group, 12 (60%) patients suffered from pain in hip straining situations.

Among the remaining 140 patients, migration and deformity of the femoral head were found to be closely related. Univariate logistic regression analysis of these combined factors showed a significant association with hip pain (odds ratio 2.79 (1.01–7.70)). Pain was also related to severe asymmetry, expressed as a difference in migration percentage in the left versus right hip. Osteoarthritis was not significantly associated with pain.

We conclude that there is a high prevalence of hip pain after unsuccessful bony hip surgery in patients with severe CP. Migration and deformity of the femoral head

are closely inter-related, and are associated with pain. Pain is also associated with asymmetry of migration of the femoral head.

Treatment should aim to prevent migration and deformity of the femoral head and to maintain as much symmetry as possible. If bony surgery is considered, the risk of residual pain due to failed surgery and/or complications is an important aspect that should be taken into consideration.

Further research could investigate changes in the configuration of the hip in CP patients after they reach adulthood.

It is not clear why 10% of patients suffer from pain in the hip region without abnormalities on the pelvic X-ray that could explain this pain. Future research with new techniques, such as the new 3D CT scan technique, but also histological investigation, might offer new insights into the factors contributing to pain in the hip joints of CP patients. Further research would be facilitated by unambiguous use of definitions, cut-off points, classification systems and assessment instruments and a sufficient duration of the follow-up.

Chapter 4 presents a study of the relationship between hip disorders (migration and deformity of the femoral head) on the one hand, and handling problems, seating problems and impairments (decubitus ulcers, fractures and contractures) on the other. This was a cross-sectional study among 160 adult patients with severe CP, the same sample used in the study described in chapter 3.

Although no significant associations were found between hip disorders and handling problems, both migration and deformity of the femoral head were positively related to the need for a special molded seat in the wheelchair, and were negatively associated with the ability to abduct the hip. Patients with seating problems had a higher mean migration percentage then patients without seating problems; the difference being significant. Also, limited passive hip abduction was significantly related to a higher mean MP in the group. Seating problems were found in 51.3% of the patients with no hip deformity, 51.8% of the patients with moderate deformity and 87.2% of the patients with severe deformity. Limited abduction was found in 39.0, 53.4 and 75.6% of patients with no, moderate and severe hip deformity, respectively.

We conclude that migration and deformity of the femoral head should be prevented in patients with severe CP. Even in the hands of experienced caregivers and therapists, people with CP GMFCS level V are difficult to handle due to their spasticity, joint

contractures, persistent reflexes etc. Further research could be aimed at facilitating the care for these patients, for instance by using antispastic drugs, administering botulinum toxin to facilitate seating, etc.

Chapter 5 presents the results of a systematic review of the effectiveness of preventive and corrective surgical interventions to treat hip luxation in patients with severe cerebral palsy. Data sources were obtained by a systematic literature search in MEDLINE, CINAHL, EMBASE, Cochrane controlled trial register and PEDRO.

We applied the following inclusion criteria: severe cerebral palsy (GMFCS levels IV and V), evidence of hip subluxation or dislocation (assessed as migration percentage (MP)), use of surgical procedure, and using decreased MP and pain relief as outcome measures. A qualitative analysis according to Steultjens et al.[1] and Van Tulder et al.[2] was performed for soft tissue surgery and osteotomies. We used the best-evidence synthesis technique to summarize the results of the included studies. All studies were observational.

Only one of the five studies involving soft tissue surgery was of sufficient quality. Nine of the ten studies involving osteotomies were of sufficient quality, including a total of 189 patients. As a result of osteotomy, the mean MP at follow-up ranged from 6 to 29%, with better results for the combination of varus derotational osteotomy (VDRO) and pelvic osteotomy than for VDRO only. No relationship could be established between the effect of the surgical procedure and the patients' age at the time of the surgery or the duration of follow-up. The percentage of patients reporting pain decreased from 81% preoperatively to 5% at follow-up, though the evidence level was insufficient. Twenty-five percent had complications like osteoarthritis, ulcers or fractures.

We conclude that there is insufficient evidence for the effectiveness of soft tissue surgery in stabilizing the hip, due to insufficient quality of the retrospective observational studies.

There are indicative findings for an effect of bony surgery in stabilizing the hip. Timing of the procedures remains an undecided issue. Multicenter trials with larger groups of patients that can be followed for a longer period could shed further light on this complex subject. We had to recalculate the mean pre- and post-operative percentages of subluxation/dislocation from a variety of articles due to different definitions of outcome measures in the articles. Further research would be facilitated by unambiguous use of definitions describing hip disorders and the use of cut-off points in, for instance, the definition of subluxation versus normal on the one hand and subluxation versus luxation on the other.

Chapter 6 presents the results of a systematic review of the effectiveness of palliative hip surgery in severe CP. Palliative intervention is undertaken when preventive or corrective surgery fails or has not been performed. A number of salvage interventions have been described.

Just as in chapter 5, we applied the following inclusion criteria: severe cerebral palsy (GMFCS IV and V), evidence of hip subluxation or dislocation (assessed as migration percentage (MP)), use of surgical procedure, and using decreased MP and pain relief as outcome measures. We performed a qualitative analysis according to Steultjens et al.[1] and Van Tulder et al.,[2] using the best-evidence synthesis technique to summarize the results of the included studies. All studies were observational.

We found articles on resection surgery of the femoral head, arthrodesis of the hip joint and total hip replacement. According to the best evidence synthesis, the indicative findings of these studies suggest that relief of pain and seating and nursing problems can be achieved after a salvage resection of the femoral head. Regrettably, none of the intervention methods (resection surgery, hip replacement or arthrodesis) provides the ultimate solution to treat the severely deformed painful hip in CP. The published literature does not clearly favor one procedure over the others. However, the technique for which the largest number of results has been published is subtrochanteric resection, which seems to eventually lead to a good outcome. Our review concludes that, if resection surgery is to be performed, distal resection and interposition surgery according to Castle and Schneider[3] should be the method of choice. The percentage of pain relief after this intervention is 90–100%. Bony intervention, however, implies no instant cure: we note that it can take up to three to six months postoperatively before maximum pain relief is obtained.

As for the technique of valgisating derotation osteotomy as described by Schanz, which is practiced by a number of orthopedic surgeons, we found no articles that could pass our quality criteria. One author reported 98% pain relief, while another described a complication percentage of 63% for this technique.

We only found one article that could pass our quality criteria that reported on other techniques like total hip replacement respectively arthrodesis of the hip joint. These techniques can produce pain relief, but also cause a relatively large number of complications. In total hip replacement, dislocation of the prosthesis occurs in 29% of the cases, heterotopic ossification (HO) is present in 45%. In addition, femoral fractures

are relatively frequent (36%). Arthrodesis of the hip joint can result in a pain-free hip, but the rate of complications, such as non-union, is high (42%).

Bony surgery can be used as a palliative intervention to relieve pain and discomfort in patients with cerebral palsy and hip disorder. Unfortunately, however, heterotopic ossification is a major problem complicating surgery in these patients. Research should be aimed at preventing this phenomenon in the future.

Chapter 7 synthesizes the results of the research described in chapters 2–4 and the reviews described in chapters 5 and 6. It also describes measurements to assess subluxation and the pathophysiology and natural history of the phenomenon. Methods of intervention are explained and their value evaluated. The question whether surgery should be performed symmetrically is discussed, as is the correct timing of surgery. The chapter also evaluates the advantages and disadvantages of surgery to prevent or treat hip disorders, and comments on the role of botulinum toxin. In addition, it discusses the different options for surveillance of hip disorders and the corresponding measures for prevention or treatment. The small hazards of frequent X-ray assessment are also discussed.

All this culminates in a proposal for a **decision tree** for the surveillance and intervention of hip disorders in spastic CP. The algorithm is based on our research and on the literature assessed in our reviews. It is now regarded as the Dutch consensus and was discussed at the general meetings of Dutch pediatric physiatrists. We compare the algorithm with the Australian consensus on surveillance, as published in 2008.

The decision tree uses the GMFCS level as a basis for intervention and is divided into three periods based on the age of the patient. The first period concerns the ages between one and four years, in which a bilateral adductor tenotomy is the preferred treatment option for most patients when an imminent hip displacement is diagnosed. The second period concerns the ages between eight and nine years, in which patients with hip disorder can be treated with VDRO, whether or not combined with pelvic osteotomy. The third period is the period after the age of nine years, in which palliative surgery can be used to treat patients with otherwise incurable pain or impairments.

We use the cerebral palsy hip classification system (CPHCS) to decide whether an intervention is needed; we think that adductor tenotomy can be useful at every GMFCS level, provided the parents are fully informed about the reason for the operation and its success rate. Bony interventions should be reserved for patients with GMFCS levels I–III, and only exceptionally used in patients with higher GMFCS levels.

Chapter 8 presents a general discussion of the research undertaken for this thesis, using the problems of a specific patient as the point of departure. The chapter addresses the main issues of the research studies and reviews we undertook, and applies them to the patient's problems. In this chapter we also put the results of our research into perspective: our patient study was only a cross-sectional and not a prospective study. Only a few articles in the reviews satisfied our quality criteria, so we had to use the best evidence method to draw conclusions.

The main conclusion is that hip disorders in spastic CP cause pain and impairments, and should if possible be prevented or treated. But not at any price: careful assessment, evaluation and discussion are necessary for each individual patient and evaluation and treatment should be performed by an experienced and patient-oriented team of professionals with affinity for the specific population.

The general discussion also includes some recommendations for future research. A suggestion is made for further research into the value of soft tissue surgery for young CP children with a high GMFCS classification. A plea is made for a properly designed system of epidemiological research into the incidence and prevalence of CP in the Netherlands and the effects of interventions, comparable to the research performed in Australia and Sweden. Such a system would make it possible, for instance, to determine the value of concentrating surgical interventions for CP children and adults in specialized centers.

In addition, further research should address the value of the common practice of passively stretching the hips and knees of CP patients, as our study shows that this is a very painful procedure for the patients.

REFERENCES

1. Steultjens EMJ, Dekker J, Bouter LM, Nes van de JCM, Lambregts BLM, Ende van den CHM. Occupational therapy for children with cerebral palsy: a systematic review. Clinical Rehab 2004;18;1-14.
2. Van Tulder MW, Assendelft WJ, Koes BW, Bouter LM. Method guidelines for systematic reviews in the Cochrane Collaboration Back Review Group for Spinal Disorders. Spine (Phila Pa 1976) 1997;22-20:2323-30.
3. Castle ME, Schneider C. Proximal femoral resection-interposition arthroplasty. J Bone Joint Surg Am 1978;60(8):1051-4.